

高雄榮民總醫院 威爾姆氏腫瘤診療原則

2019年01月21日 第一版

兒童癌症醫療團隊擬訂

注意事項：這個診療原則主要作為醫師和其他保健專家診療癌症病人參考之用。假如你是一個癌症病人，直接引用這個診療原則並不恰當，只有你的醫師才能決定給你最恰當的治療。

修訂指引

- 本共識依下列參考資料制定版本
– 台灣兒童癌症研究群(TPOG)，
TPOG_WT_ V2_20170327_

會議討論

上次會議：2018/02/22

本共識與上一版的差異

上一版	新版
<ul style="list-style-type: none">1. 無治療流程圖。2. 無化學治療處方建議總表。	<ul style="list-style-type: none">1. 新增治療流程圖。2. 新增化學治療處方建議總表。

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

腫瘤分級

Stage	COG (pre-chemotherapy)	SIOP (post-chemotherapy)
I	Tumor is limited to kidney and is completely resected	Tumor limited to kidney or surrounded with fibrous pseudocapsule if outside the normal contour of the kidney, the renal capsule or pseudocapsule may be infiltrated with the tumor, but it does not reach the outer surface, and it is completely resected (resection margins "clear")
	Renal capsule intact, not penetrated by tumor	The tumor may be protruding (bulging) into the pelvic system and "dipping" into the ureter, but it is not infiltrating their walls
	No tumor invasion of veins or lymphatics of renal sinus	The vessels of the renal sinus are not involved, but intrarenal vessel involvement may be present.
	No nodal or hematogenous metastases	Fine needle aspiration or percutaneous core needle biopsy ("tru-cut") do not upstage the tumor. The presence of necrotic tumor or chemotherapy-induced changes in the renal sinus/hilus fat and/or outside of the kidney should not be regarded as a reason for upstaging a tumor
	No prior biopsy	
	Negative margins	
II	Tumor extends beyond kidney but completely resected	The tumor extends beyond kidney or penetrates through the renal capsule and/or fibrous pseudocapsule into perirenal fat but is completely resected (resection margins "clear")
	Tumor penetrates renal capsule	The tumor infiltrates the renal sinus and/or invades blood and lymphatic vessels outside the renal parenchyma but it is completely resected
	Tumor in lymphatics or veins of renal sinus	The tumor infiltrates adjacent organs or vena cave but is completely resected
	Tumor in renal vein with margin not involved	
	No nodal or hematogenous metastases	
	Negative margin	

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

腫瘤分級

III	Residual tumor or nonhematogenous metastases confined to abdomen	Incomplete excision of the tumor which extends beyond resection margins (gross or microscopic tumor remains postoperatively)
	Involved abdominal nodes	Any abdominal lymph nodes are involved
	Peritoneal contamination or tumor implant	Tumor rupture before or intraoperatively (irrespective of other criteria for staging)
	Tumor spillage of any degree occurring before or during surgery	The tumor has penetrated through the peritoneal surface
	Gross residual tumor in abdomen	Tumor implants are found on the peritoneal surface
	Biopsy of tumor (including fine needle aspiration) prior to removal of kidney	The tumor thrombi present at resection margins of vessels or ureter, transected or removed piecemeal by surgeon
	Resection margins involved by tumor or transection of tumor during resection (i.e. piecemeal excision of tumor)	The tumor has been surgically biopsied (wedge biopsy) prior to preoperative chemotherapy or surgery
		The presence of necrotic tumor or chemotherapy-induced changes in a lymph node or at the resection margins should be regarded as stage III
IV	Hematogenous metastases or spread beyond abdomen	Hematogenous metastases or spread beyond abdomen
V	Bilateral renal tumors Each side's tumor should be substaged separately according to the above criteria	Bilateral renal tumors Each side's tumor should be substaged separately according to the above criteria

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

化學治療處方建議表

Regimen	Agents
EE4A	vincristine and dactinomycin
DD4A	vincristine, dactinomycin, doxorubicin and possibly radiation therapy
I	vincristine, dactinomycin, doxorubicin, cyclophosphamide, and etoposide
M	vincristine, dactinomycin, doxorubicin, cyclophosphamide, and etoposide
rUH-1	vincristine, dactinomycin, doxorubicin, cyclophosphamide, carboplatin, and etoposide
UH2	vincristine, dactinomycin, doxorubicin, cyclophosphamide, carboplatin, etoposide, and irinotecan
vincristine/irinotecan window therapy	vincristine and irinotecan in conjunction with revised UH-1 or UH-2 depending on response

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

單側威爾姆氏種類危險級分類

Patient age	Tumor weight	Stage, histology	LOH at both 1p and 16q	Rapid lung nodule response	Chemotherapy regimen	Radiation therapy
< 2 yrs	< 550 g	I, FH	Any	N/A	None	None
< 2 yrs	≥ 550 g	I, FH	None	N/A	EE4A	None
> 2 yrs	Any	I, FH	None	N/A	EE4A	None
Any	Any	II, FH	None	N/A	EE4A	None
Any	Any	I, FH	LOH*	N/A	DD4A	None
Any	Any	II, FH	LOH*	N/A	DD4A	None
Any	Any	III, FH	None	Any	DD4A	Local
Any	Any	III, FH	LOH*	Any	M	Local
Any	Any	IV, FH	None	Yes	DD4A	Local
Any	Any	IV, FH	None	No	M	Local, lung
Any	Any	IV, FH	LOH*	Any	M	Local, lung
Any	Any	I, FA or DA	Any	Any	DD4A	Local
Any	Any	II-III, FA	Any	Any	DD4A	Local
Any	Any	I-III CCSK	Any	Any	I	Local (RT omitted for stage I)
Any	Any	II-III, DA	Any	Any	rUH-1	Local
Any	Any	IV CCSK	Any	Any	rUH-1	Local
Any	Any	I-IV RTK	Any	Any	RTK	Local
Any	Any	IV, FA or DA	Any	Any	UH-2	Local, lung

This risk-stratification schema was modified from an existing risk stratification (Jeffrey et al 2014) and COG AREN0534.

*The detection of LOH at chromosomes 1p and 16q is optional if not applicable.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

治療原則及療程表表示方法

1. 若能手術切除，以nephrectomy為主要優先治療(並盡量不要經皮切片診斷)；若無法手術切除，先進行neo-adjuvant chemotherapy
2. Nephrectomy: on day 0 of week 0.
3. For “biopsy only” patient, definitive surgery is undertaken at week 7 or week 13 after preoperative chemotherapy.
4. Chemotherapy should be administered within 14 days post-nephrectomy.
5. Week 1 = day 7 post nephrectomy.
6. Newborns and all <12 months old require a reduction in chemotherapy doses to 50% of those given to older children.
7. RT: over 5-7 days after nephrectomy

手術前化學治療

符合下列條件者，考慮先進行neo-adjuvant chemotherapy：

1. Synchronous bilateral Wilms tumor
2. Wilms tumor in a solitary kidney
3. Extension of tumor thrombus in the inferior vena cava above the level of the hepatic veins
4. Tumor involved contiguous structures whereby the only means of removing the kidney tumor requires removal of the other structures (e.g. spleen, pancreas, or colon but excluding the adrenal gland).
5. Inoperable Wilms tumor
6. Pulmonary compromise due to extensive pulmonary metastases

兒癌-威爾姆氏腫瘤

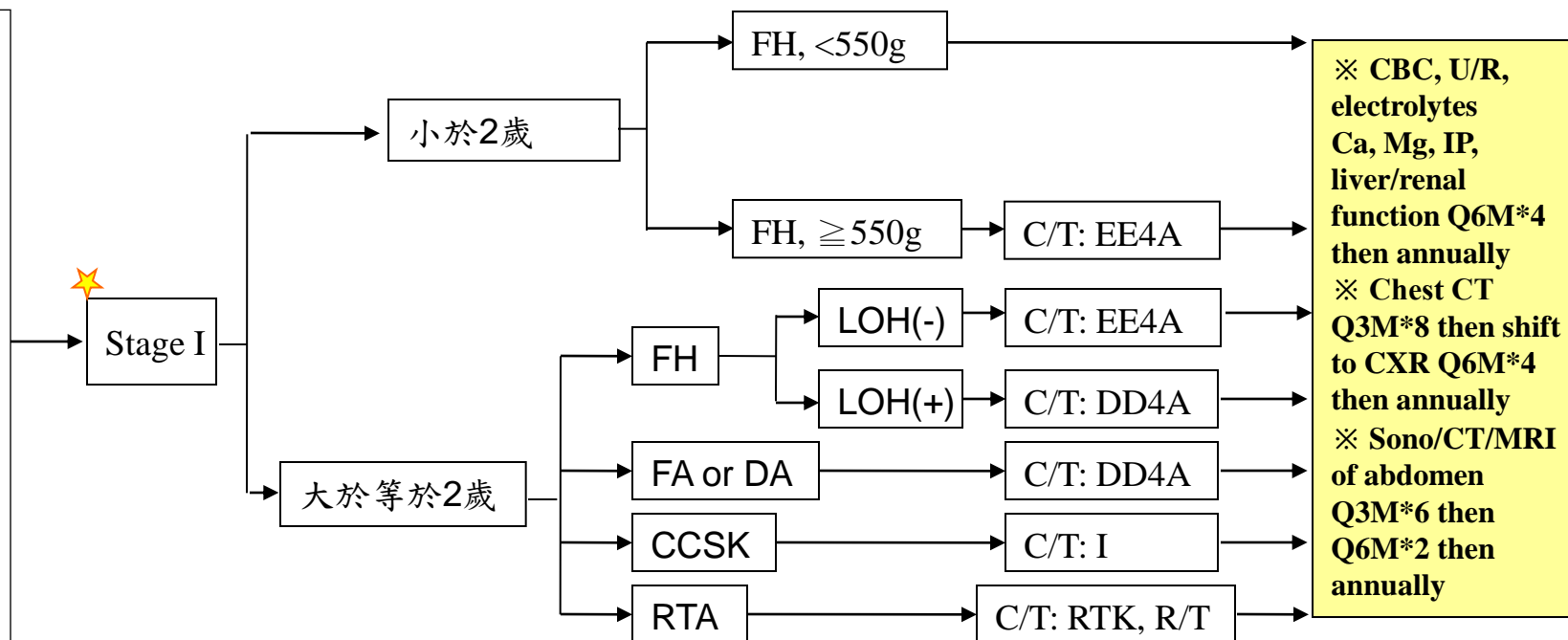
高雄榮民總醫院
臨床診療指引

2019年第一版

評估	診斷	治療	追蹤
----	----	----	----

- 病史，理學檢查
- 營養及日常體能狀態
- 身高體重，體表面積計算
- 血液常規
- 電解質及肝腎功能
- 凝血功能
- 心臟超音波檢查
- 腹部超音波
- 聽力檢查
- 腫瘤病理種類*
- 骨頭掃描*
- 胸部電腦斷層(CT)*
- 腹部電腦斷層攝影(CT)* or 核磁共振檢查(MRI)*(擇一)
- 腫瘤之LOH染色體檢測(外送臺大醫院)*

*與癌症期別相關之主要檢查



★ 以COG stage I (pre-chemotherapy)為主，可手術完全切除且無先施行切片

兒癌-威爾姆氏腫瘤

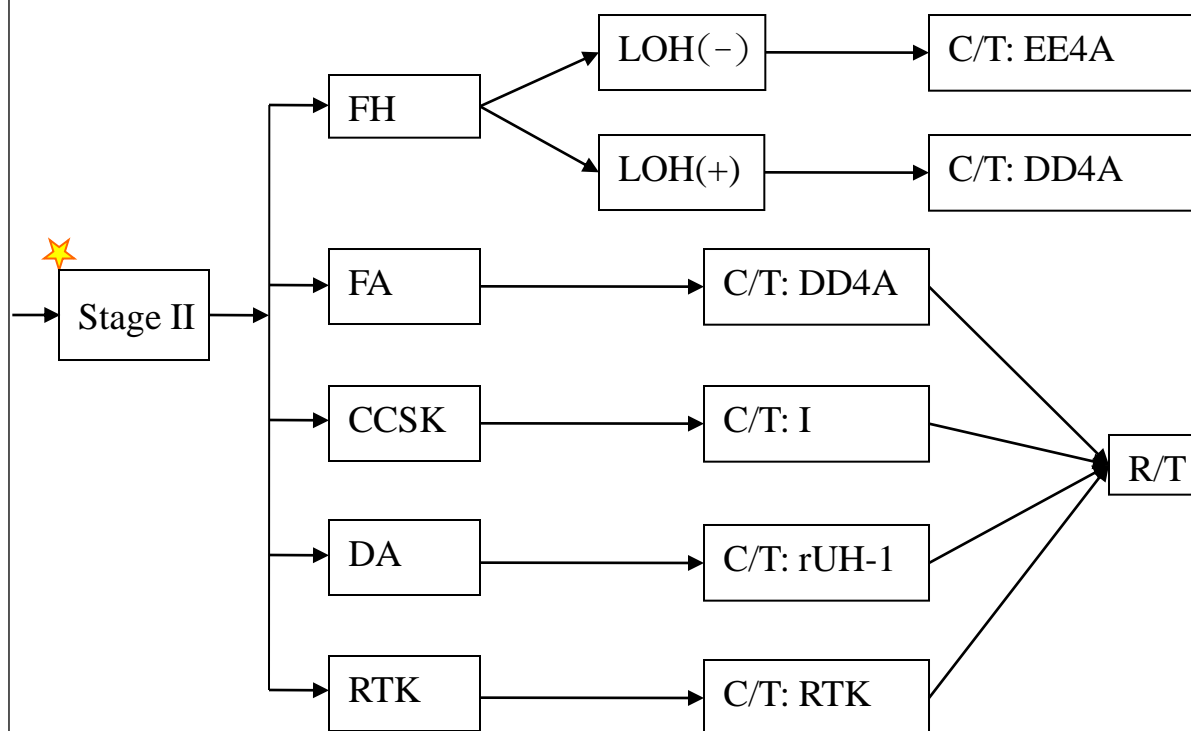
高雄榮民總醫院
臨床診療指引

2019年第一版

評估	診斷	治療	追蹤
----	----	----	----

- 病史，理學檢查
- 營養及日常體能狀態
- 身高體重，體表面積計算
- 血液常規
- 電解質及肝腎功能
- 凝血功能
- 心臟超音波檢查
- 腹部超音波
- 聽力檢查
- 腫瘤病理種類*
- 骨頭掃描*
- 胸部電腦斷層(CT)*
- 腹部電腦斷層攝影(CT)* or 核磁共振檢查(MRI)*(擇一)
- 腫瘤之LOH染色體檢測(外送臺大醫院)*

*與癌症期別相關之主要檢查



※ CBC, U/R, electrolytes Ca, Mg, IP, liver/renal function Q6M*4 then annually
 ※ Chest CT Q3M*8 then shift to CXR Q6M*4 then annually
 ※ Sono/CT/MRI of abdomen Q3M*6 then Q6M*2 then annually

★ 以COG stage II (pre-chemotherapy)為主

兒癌-威爾姆氏腫瘤

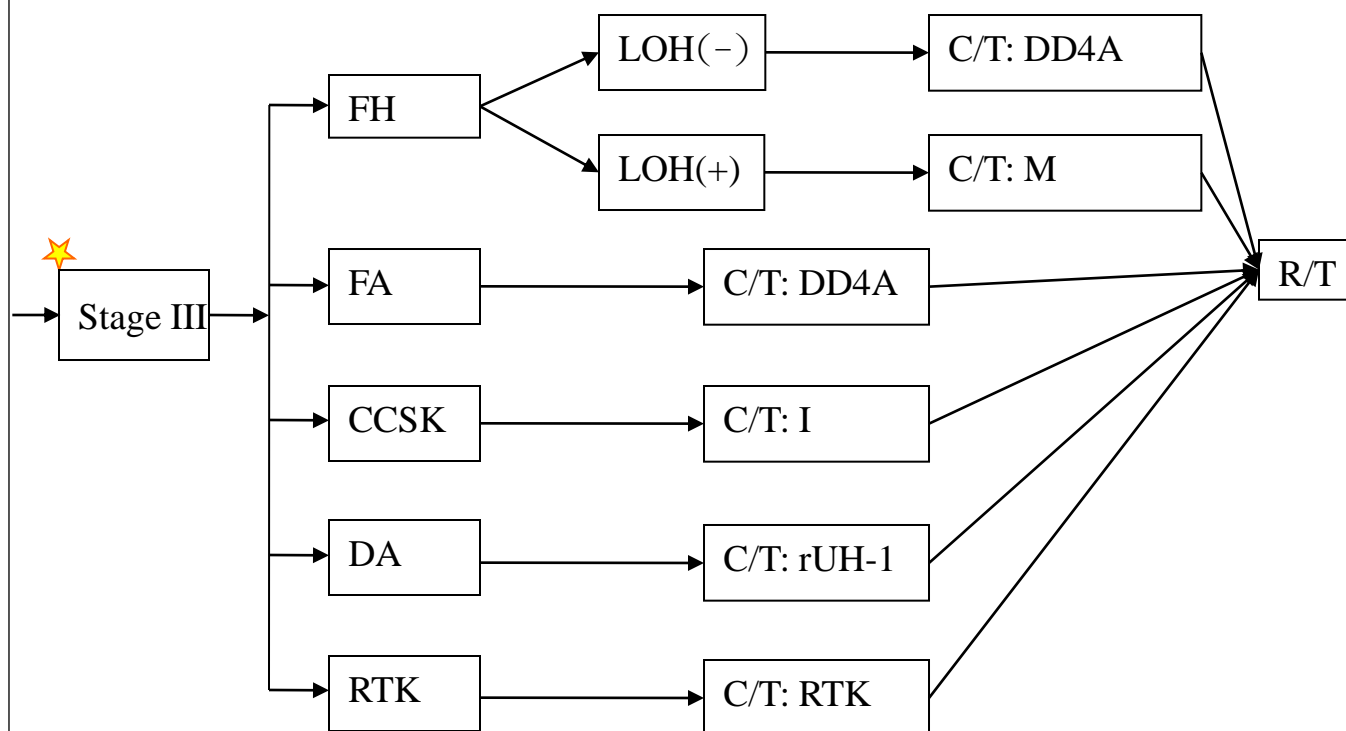
高雄榮民總醫院
臨床診療指引

2019年第一版

評估	診斷	治療	追蹤
----	----	----	----

- 病史，理學檢查
- 營養及日常體能狀態
- 身高體重，體表面積計算
- 血液常規
- 電解質及肝腎功能
- 凝血功能
- 心臟超音波檢查
- 腹部超音波
- 聽力檢查
- 腫瘤病理種類*
- 骨頭掃描*
- 胸部電腦斷層(CT)*
- 腹部電腦斷層攝影(CT)* or 核磁共振檢查(MRI)*(擇一)
- 腫瘤之LOH染色體檢測(外送臺大醫院)*

*與癌症期別相關之主要檢查



※ CBC, U/R, electrolytes
Ca, Mg, IP, liver/renal function Q6M*4 then annually
※ Chest CT Q3M*8 then shift to CXR Q6M*4 then annually
※ Sono/CT/MRI of abdomen Q3M*6 then Q6M*2 then annually

★ 以COG stage III (pre-chemotherapy)為主

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

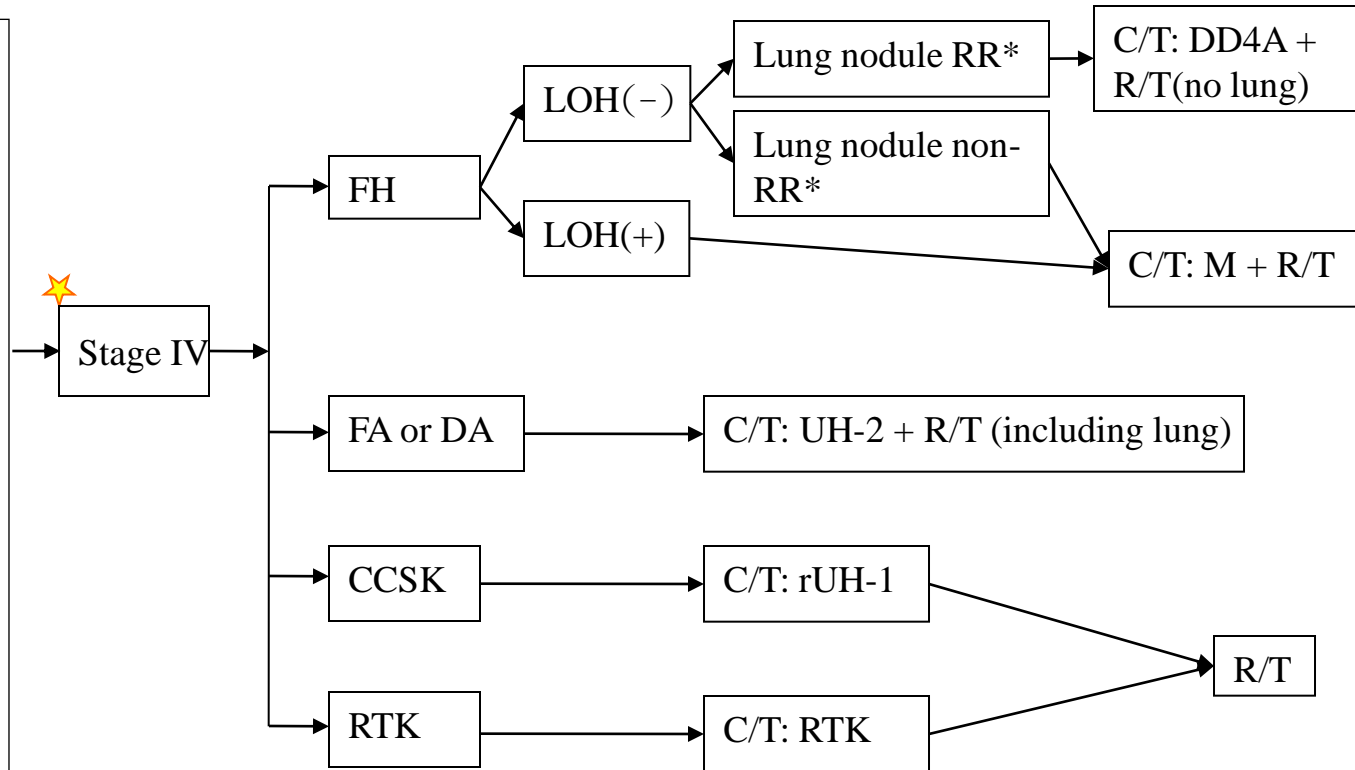
評估

診斷

治療

追蹤

- 病史，理學檢查
- 營養及日常體能狀態
- 身高體重，體表面積計算
- 血液常規
- 電解質及肝腎功能
- 凝血功能
- 心臟超音波檢查
- 腹部超音波
- 聽力檢查
- 腫瘤病理種類*
- 骨頭掃描*
- 胸部電腦斷層(CT)*
- 腹部電腦斷層攝影(CT)* or 核磁共振檢查(MRI)*(擇一)
- 腫瘤之LOH染色體檢測(外送臺大醫院)*



※ CBC, U/R, electrolytes
Ca, Mg, IP, liver/renal function Q6M*4 then annually
※ Chest CT Q3M*8 then shift to CXR Q6M*4 then annually
※ Sono/CT/MRI of abdomen Q3M*6 then Q6M*2 then annually

*與癌症期別相關之主要檢查

★以COG stage IV (pre-chemotherapy)為主
**RR: rapid response

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

手術前化學治療處方建議表 Regimen VAD

Stage I-IV bilateral Wilms tumor (BWT) with biopsy revealing favorable histology or no preoperative biopsy ; stage I-III BWT with focal anaplasia ; stage I BWT with diffuse anaplasia ; or high-risk, stage III-IV unilateral Wilms tumor with contralateral nephrogenic rest or predisposition syndrome

WK	1	2	3	4	5	6
VCR	v	v	v	v	v	v
AMD	v			v		
EPI	v			v		

Vincristine (VCR): 0.05 mg/kg IV push if BW is < 30 kg; 1.5 mg/M² IV push if BW is > 30kg (maximal dose 2 mg) weeks 1 to 6.

Dactinomycin (AMD): 0.045mg/kg IV push over 5 minutes x 1 dose, 1.35 mg/M² if BW is >30kg (maximal single dose 2.3 mg) on weeks 1 and 4.

Epirubicin (EPI): 1.5mg/kg IV infusion over 1-2 hours, 45 mg/M² if BW is >30kg on week 1 and 4.

** Calculating drug dosage on the basis of surface area probably leads to an overestimation in infants, so doses usually are calculated according to body weight instead. However, the absence of any other severe acute toxicities (particularly neutropenia) in the current series may indicate that a 50% reduction of the dose calculated by body weight may be excessive. One option is to recommend a 33% dose reduction for children age < 6 months.

** Reduction of all drugs for infants to 2/3 of the doses for older children. (infant除了用kg算之外，還要再減少 1/3 的劑量)

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

化學治療處方建議表 Regimen EE4A

Stage I / FH and stage II / FH: Nephrectomy, chemotherapy using Regimen EE4A

↓ reevaluate

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19
VCR	v	v	v	v	v	v	v	v	v	v			V*			V*			V*
AMD	v			v			v			v			v			v			v

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2 mg), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established; then weekly for a total of 10 doses. The dose of vincristine is 1.5 mg/M² IV push for all patients who weigh more than 30 kg, but no single dose should exceed 2.0 mg.

Vincristine (VCR, V*) 0.067 mg/kg IV push (maximum dose - 2.0 mg) with dactinomycin at weeks 13, 16 and 19. The dose of vincristine is 2.0 mg/M² IV push with dactinomycin for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Dactinomycin (AMD) 0.045 mg/kg/dose IV push (maximum dose - 2.3 mg), beginning within 7 days post-nephrectomy (during week 0), and then at weeks 4, 7, 10, 13, 16, and 19. The dose of dactinomycin is 1.35 mg/M² for all patients who weighed more than 30 kg, but no single dose should exceed 2.3 mg.

Chemotherapy guidelines (Note: The day of nephrectomy will be considered day 0; the first dose of chemotherapy will be measured in days from that starting point.) No dose of dactinomycin should be initiated if the absolute neutrophil count is <1,000/mm³ or the platelet count is <100,000/mm³.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

化學治療處方建議表 Regimen DD4A

Stage III / FH; Stage I / Focal or diffuse anaplasia; Stage II or III / Focal anaplasia:
Nephrectomy, abdominal irradiation, chemotherapy using Regimen DD-4A

↓ reevaluate

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25
VCR	v	v	v	v	v	v	v	v	v	v			V*			V*			V*			V*			V*
AMD	v						v						v						v						v
EPI				v						v						V*						V*			

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2 mg), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established, then weekly for a total of 10 doses. The dose of vincristine is 1.5 mg/M² IV push for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Vincristine (VCR, V*) 0.067 mg/kg IV push (maximum dose - 2 mg) with dactinomycin or epirubicin at weeks 13, 16, 19, 22 and 25. The dose of vincristine is 2.0 mg/M² IV push for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Dactinomycin (AMD) 0.045 mg/kg/dose IV push (maximum dose - 2.3 mg), beginning within 7 days post-nephrectomy (during week 0), and then at weeks 7, 13, 19, and 25. The dose of dactinomycin administered at week 7 should be decreased by 50% (0.0225 mg/kg/dose) if whole lung or whole abdomen radiation therapy has been given. The dose of dactinomycin is 1.35 mg/M² for all patients who weighed more than 30 kg, but no single dose should exceed 2.3 mg. The dose of dactinomycin administered at week 7 should be decreased by 50% (0.675 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

Epirubicin (EPI) 1.5 mg/kg IV infusion over 1-2hours, is given at weeks 4 and 10; subsequently, 1.0 mg/kg IV push is given at weeks 16 and 22. The dose of epirubicin administered at week 3 should be decreased by 50% (0.75 mg/kg) if whole lung or whole abdomen radiation therapy has been given. The dose of epirubicin at weeks 4 and 10 is 45 mg/M² IV push, and at weeks 16 and 22 is 30 mg/M² IV push for all patients who weighed more than 30 kg. The dose at week 4 should be decreased by 50% (22.5 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

化學治療處方建議表 Regimen M (modified DD4A)

- Chest CT will be performed on all Stage IV patients with lung metastases at study enrollment and at Week 6.
- Patients who have complete disappearance of their lung metastases (or who have tissue confirmation that the nodules do not contain viable tumor) at the Week 6 evaluation will be considered rapid responders and will continue with DD-4A.
- Patients who do not have complete resolution of pulmonary nodules by Chest CT will undergo pulmonary irradiation and will be switched to regimen M (DD4A variation with dactinomycin and epirubicin given on the same day and alternating cyclophosphamide and etoposide)

WK	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
VCR		v	v		v	v	v			V*						V*					V*				V*
CTX ³	v			v								v						v							
VP-16 ³	v			v								v						v							
AMD							v			v						v					v				v
EPI							v			v						v					v				v

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

化學治療處方建議表 Regimen M (modified DD4A)

Vincristine (VCR) 0.05mg/kg ($1.5\text{mg}/\text{M}^2$ if BW >30kg), iv push at weeks 8, 9, 11, 12, 13.
Maximal single dose is 2mg.

Vincristine (VCR, V*) 0.067mg/kg ($2\text{mg}/\text{M}^2$ if BW >30kg) at weeks 16, 22, 28, 31. Maximal single dose is 2mg.

Cyclophosphamide⁵ (CTX⁵) and Mesna with Etoposide (VP-16⁵): days 1-5, at weeks 7, 10, 19, 25.

Administration schedule:

-2 to 0 hr: Hydration at a rate of $200\text{ml}/\text{M}^2$ /hr for 2 hours with D5 1/4 NS, IVF.

0 to 1 hr: CTX⁵ $14.7\text{mg}/\text{kg}$ ($440\text{mg}/\text{M}^2$ if BW >30kg) + Mesna $3\text{mg}/\text{kg}$ in $200\text{ml}/\text{M}^2$ D5 1/2 NS IV infusion for 1 hour.

1-2 hr: VP-16⁵ $3.3\text{mg}/\text{kg}$ in $200\text{ml NS}/\text{M}^2$ IV over 1 hr ($100\text{mg}/\text{M}^2$ if BW >30 kg)

3, 6, 9 hr: Mesna $3\text{mg}/\text{kg}$ (or $90\text{mg}/\text{M}^2$ if BW >30kg) in 10ml NS IV infusion 15 min., q3h for 3 doses. Continue hydration at $150\text{ml}/\text{M}^2$ /hr for 6 hours with D5 1/2 NS

9-22 hr: D5 1/2 NS at $1000\text{ml}/\text{M}^2$ (total)

22-23 hr: same as -2 to 0 hrs.

Dactinomycin (AMD): $0.045\text{mg}/\text{kg}$ per dose, IV over 15 minutes. ($1.35\text{mg}/\text{M}^2$ /dose if BW > 30 kg), (maximal single dose 2.3 mg) at weeks 13, 16, 22, 28, 31. Consider dose reduce by 50% at week 16, if delayed RT has been feasible at week 13.

Epirubicin (EPI): $1\text{mg}/\text{kg}$ IV in $200\text{ml}/\text{M}^2$ D5 1/2 NS, IV infusuin over 1-2 hours ($30\text{mg}/\text{M}^2$ if BW >30 kg) at weeks 13, 16, 22, 28, 31. Dose should be reduced by 50% at week 16 if delayed RT has been feasible at week 13.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

化學治療處方建議表 Regimen I

Stage I-III / Clear cell sarcoma of the kidney(CCSK) : Nephrectomy, abdominal irradiation using 1080 cGy for Stage II & III patients, whole lung irradiation for patients with pulmonary metastases, chemotherapy with vincristine, epirubicin, etoposide, cyclophosphamide and mesna using Regimen I (see below).

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25
VCR	v	v	v		v	v	v	v	v		v	v	V*	V*					V*						V*
EPI	v						v						v						v						v
CTX ³							v						v						v						v
CTX ³				v						v						v						v			
VP-16 ³				v						v						v						v			

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2mg.), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established, then at weeks 1-3, 5-9, 11-12. The dose of VCR is 1.5 mg/M² IV push for all patients who weighed more than 30 kg., but no single dose should exceed 2.0 mg.

Vincristine (VCR,V*) 0.067 mg/kg IV push (maximum dose - 2 mg) at week 13, 14, 19, 25. The dose of VCR is 2.0 mg/M² IV push for all patients who weighed more than 30 kg., but no single dose should exceed 2.0 mg.

Epirubicin (EPI) 1.5 mg/kg **IV infusion over 1-2 hours**, is given at weeks 1, 7, 13, 19 and 25. The dose of EPI administered at week 7 should be decreased by 50% (0.75 mg/kg) if whole lung or whole abdomen radiation therapy has been given. The dose of EPI at weeks 1, 7, 13, 19 and 25 is 45 mg/M² IV push for all patients who weighed more than 30 kg. The dose at week 7 should be decreased by 50% (22.5 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

化學治療處方建議表 Regimen I

Cyclophosphamide (CTX³) and Mesna on days 1-3, with Epirubicin (EPI) day 1, at weeks 7, 13, 19, 25.

Administration schedule:

-2 to 0 hr: Hydration at a rate of 200 ml/M² /hr for 2 hours with D5 1/2 NS IVF.

0 to 1 hr: CTX³ 14.7 mg/kg (440 mg/M² if BW >30kg) + Mesna 3 mg/kg in 200 ml/M² D5/ 1/2 NS IV infusion for 1 hour.

1-2 hr: EPI: 1.5mg/kg IV in 200 ml/M² D5 1/2 NS, iv over 1-2 hours (45 mg/M² if BW >30kg) at day 1

if RT has been given, or at week 19 if delayed tumor resection and RT is feasible at week 13.

3, 6, 9 hr: Mesna 3 mg/kg (or 90 mg/M² if BW >30kg) in 10ml NS iv infusion 15 min., q3h for 3 doses on days 1-3. Continue hydration at 150 ml/M²/hr for 6 hours with D5 1/2 NS.

9-22 hr: D5 1/2 NS at 1000 ml/M² (total)

22-23 hr: same as -2 to 0 hrs.

Cyclophosphamide (CTX⁵), and Mesna with Etoposide(VP-16⁵) on days 1-5, at weeks 4, 10, 16, 22.

Administration schedule:

-2 to 0 hr: Hydration at a rate of 200ml/M² /hr for 2 hours with D5 1/4 NS, IVF.

0 to 1 hr: CTX⁵ 14.7 mg/kg (440mg/M² if BW >30kg) + Mesna 3mg/kg in 200ml/M² D5 1/2 NS IV infusion for 1 hour.

1-2 hr: VP-16⁵ 3.3 mg/kg in 200 ml NS /M² IV over 1 hr (100 mg/M² if BW >30 kg)

3, 6, 9 hr: Mesna 3 mg/kg (or 90 mg/M² if BW >30kg) in 10ml NS IV infusion 15 min., q3h for 3 doses on days 1-5. Continue hydration at 150 ml/M² for 6 hours with D5 1/2 NS

9-22 hr: D5 1/2 NS at 1000 ml/M² (total).

22-23 hr: same as -2 to 0 hrs.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

化學治療處方建議表 Regimen RTK

Stage I-IV / Rhabdoid tumor of the kidney: Nephrectomy, radiation therapy and chemotherapy with cyclophosphamide, mesna, etoposide and carboplatin

Babies <12 months of age should receive ONE-HALF of the recommended doses of all chemotherapeutic agents, as calculated on the basis of body weight. Full doses of chemotherapeutic agents should be administered to those patients when the child is ≥ 12 months of age.

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25
CBP ²	v			v						v			v						v			v			
VP-16 ³	v			v						v			v						v			v			
CTX ⁴							v									v									v

Carboplatin (CBP²) 16.7 mg/kg/day x 2 days, IV infusion over 60 minutes at weeks 1, 4, 10, 13, 19, 22. The dose of carboplatin is 500 mg/M²/day x 2 days for all patients who weighed more than 30 kg.

Etoposide (VP-16³) 3.3 mg/kg/day x 3 days in 200 ml/M² of D5 1/2 NS as an IV infusion over 60 minutes daily is given at weeks 1, 4, 10, 13, 19, 22 after carboplatin infusion. The dose of etoposide is 100 mg/M²/day x 3 days for all patients who weighed more than 30 kg.

Cyclophosphamide (CTX⁴) 14.7 mg/kg/day x 4 days (or 5 days) in 200 ml/M² of D5 1/2 NS as an IV infusion over 60 minutes daily is given at weeks 7, 16, 25. The dose of cyclophosphamide is 440 mg/M²/day x 5 days for all patients who weighed more than 30 kg.

Mesna 3 mg/kg/dose x 4 doses in 10 ml IV over 15 minutes x 5 days, given after cyclophosphamide, at weeks 7, 16, and 25. The dose of mesna should be 90 mg/M²/dose x 4 doses x 5 days for all patients who weighed more than 30 kg.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年第一版

放射治療處方建議表

	Histology				
	Favorable	Focal anaplasia	Diffuse anaplasia	Clear cell sarcoma	Rhabdoid tumor
Stage I	0	10.8	10.8	10.8	10.8/19.8*
II	0	10.8	10.8	10.8	10.8/19.8*
III	10.8	10.8	<u>19.8</u>	10.8	10.8/19.8*
IV	Based on abdominal stage and histology				

* For patients aged > 1 year old

1. All patients except stage I and II favorable histology are irradiated.
2. Focal and diffuse anaplasia are distinguished in that Stage III diffuse anaplasia gets a higher dose
3. Whole lung RT is given only if pulmonary nodules are not in CR after week 6.
4. RT should start by day 10 post-op and not later than day 14.
5. Boost small areas of gross lung metastasis to 20 Gy.

兒癌-威爾姆氏腫瘤

高雄榮民總醫院
臨床診療指引

2019年 第一版

癌症藥物停藥準則

影像學檢查，若治療期間腫瘤有變大、轉移情況，或有嚴重藥物毒性出現，應停止或改變治療方式。

Reference

1. Scott RH, Stiller CA, Walker L, et al. Syndromes and constitutional chromosomal abnormalities associated with Wilms tumour. *J Med Genet.* 2006;43:705-715.
2. Green DM, Breslow NE, Beckwith JB, et al. Treatment with nephrectomy only for small, stage I/favorable histology Wilms tumor: a report from the National Wilms Tumor Study Group. *J Clin Oncol.* 2001;37:19-3724.
3. Oue T. New risk classification is necessary in the treatment of Wilms tumor. *Transl Pediatr* 2014;3:39-41.
4. Zugor V1, Schott GE, Lausen B, et al. Clinical and surgical experience with Wilms' tumor. Long-term results of a single institution. *Anticancer Res.* 2010;30:1735-1739.
5. Kalapurakal JA, Li SM, Breslow NE, et al. Intraoperative spillage of favorable histology wilms tumor cells: influence of irradiation and chemotherapy regimens on abdominal recurrence. A report from the National Wilms Tumor Study Group. *Int J Radiat Oncol Biol Phys.* 2010;76:201-206.
6. Green DM, Breslow NE, D'Angio GJ, et al. Outcome of patients with Stage II/favorable histology Wilms tumor with and without local tumor spill: a report from the National Wilms Tumor Study Group. *Pediatr Blood Cancer.* 2014;61:134-139.
7. Green DM, Breslow NE, Beckwith JB, et al. Treatment outcomes in patients less than 2 years of age with small, stage I, favorable-histology Wilms' tumors: a report from the National Wilms' Tumor Study. *J Clin Oncol.* 1993;11:91-95.
8. Warmann SW1, Furtwängler R, Blumenstock G, et al. Tumor biology influences the prognosis of nephroblastoma patients with primary pulmonary metastases: results from SIOP 93-01/GPOH and SIOP 2001/GPOH. *Ann Surg.* 2011;254:155-162.
9. Grundy PE, Breslow NE, Li S, et al. Loss of heterozygosity for chromosomes 1p and 16q is an adverse prognostic factor in favorable-histology Wilms tumor: a report from the National Wilms Tumor Study Group. *J Clin Oncol.* 2005;23:7312-7321.
10. Que T. New risk classification is necessary in the treatment of Wilms tumor. *Transl Pediatr.* 2014;3:39-41.
11. Kieran K, Anderson JR, Dome JS, et al. Lymph node involvement in Wilms tumor: results from National Wilms Tumor Studies 4 and 5. *J Pediatr Surg.* 2012;47:700-706.
12. Lall A, Pritchard-Jones K, Walker J, et al. Wilms' tumor with intracaval thrombus in the UK Children's Cancer Study Group UKW3 trial. *J Pediatr Surg.* 2006;41:382-387.